Primary Malignant Fibrous Histiocytoma of Breast: A Common Tumour in an Uncommon Location

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Abstract: This report concerns a case of primary de-novo origin malignant fibrous histiocytoma of breast in a 32 years young lady involving almost the entire right breast. Fine needle aspiration cytology of the breast mass suggested a malignant spindle cell lesion. Then she was treated by simple mastectomy followed by chest wall irradiation. Histopathology together with immunohistochemistry confirmed the lesion as malignant fibrous histiocytoma, a rare de-novo origin breast neoplasm. Her post operative period was uneventful except radiation induced brachial plexopathy.

Keywords: malignant fibrous histiocytoma, breast, de-novo, rare.

1. Introduction

Primary sarcomas of breast are malignant tumours arising from the mesenchymal tissue of the breast. Primary breast sarcoma is a rare entity having frequency of <1% of all breast malignancy[1]-[2]. Pure sarcomas could be angiosarcoma, liposarcoma, leiomyosarcoma, fibrosarcoma, rhabdomyosarcoma or malignant fibrous histiocytoma(MFH). Among all these angiosarcoma is the commonest while only 30 cases of primary MFH of female breast have still been reported in literature[2]-[5]. Breast sarcomas mostly appear in the fourth and fifth decade of life. Mean age is around 40 years[6]. Many breast sarcomas arise frequently after radiotherapy for breast carcinoma[7]. We hereby report an unique case of de-novo origin primary MFH of breast in a 32 years old young lady.

2. Case Report

The present case was a female aged 32 years who presented with a rapidly growing painless right breast lump in upper outer quadrant. There was no familial history of such lesion. The patient denied any co morbidity and did not take any regular medication for any other disease.

On physical examination, almost whole breast was seemed to be involved by the hard, irregular mass. The mass was fixed to skin but not to the chest wall. No cutaneous change or axillary lymphadenopathy was noted. Ultrasound showed a solid but relatively circumscribed lesion. Her systemic examination was unremarkable and routine haematological and biochemical tests like haemogram, urea, creatinine, blood sugar were within normal limits.

Fine needle aspiration cytology(FNAC) showed low cellular smears comprising of clusters as well as singly dispersed pleomorphic cells. Individual cells had moderate to plenty amount of cytoplasm which was vacuolated at places. The nucleus was plump to spindle with irregular distribution of chromatin and prominent nucleolus (figure 1). On the basis of the above findings a provisional diagnosis of malignant spindle cell lesion was made.

Figure 1: Microphotograph of the FNAC smear showing pleomorphic spindle cells with prominent nucleolus.

Depending upon the provisional diagnosis a simple mastectomy was done and the specimen was sent to the department of pathology for histopathological diagnosis.

Grossly, there was a well circumscribed, hard mass measuring 20cm×11cm×18cm involving the whole of the breast. Outer surface was unremarkable except the fact that areola, nipple could not be identified properly (figure 2a). Cut surface was solid and greyish-white in colour having multiple whorled structures with foci of haemorrhage & necrosis (figure 2b). During grossing one section per one centimetre of the mass was given for tissue processing and histopathological examination.
Figure 2a: Areola and nipple could not be identified on the external surface of the mastectomy specimen

Figure 2b: Cut surface of the solid mass showing whorling with foci of haemorrhage and necrosis.

Microscopical examination of the haematoxylin and eosin stained slides showed sheets and fascicles of pleomorphic oval to spindle cells with storiform pattern and tumour giant cells at places (figure 3a & 3b). Mitoses were frequent throughout the tumour (average 2-3/10 high power fields). Among them many were atypical in nature. Necrosis was also noted but ductular or epithelial elements were not seen in the mass.

Figure 3a: Microphotograph showing storiform arrangement of the pleomorphic oval to spindle cells

Figure 3b: Microphotograph showing tumour giant cells and atypical mitoses

On immune histochemical staining, the tumor cells were diffusely positive for vimentin and negative for cytokeratin supporting a mesenchymal origin of the tumour. (figure 4a & 4b)

Figure 4a: Microphotograph showing tumour cells are diffusely positive for vimentin

Figure 4b: Microphotograph showing tumour cells are diffusely negative for cytokeratin

For further categorisation desmin, smooth muscle actin & CD68 were used. Among these three only CD68 showed
feature is a large number of tumor cells in various area. Third is inflammatory type where the most prominent cells, particularly neutrophilic granulocytes. Fourth is the differentiations mixed with a large number of inflammatory breast and female genital tract even does not include MFH in fascicle on the classification of malignant tumors of the uncommon site for this type of tumor. The most recent WHO upper extremities and retroperitoneum but breast is very frequently in the deep fascia or skeletal muscle of the lower extremities. Malignant fibrous histiocytoma (MFH) is a soft tissue neoplasm representing the most common soft tissue sarcoma of middle and late adulthood [8]. This tumor occurs most frequently in the deep fascia or skeletal muscle of the lower and upper extremities and retroperitoneum but breast is very uncommon site for this type of tumor. The most recent WHO fascicle on the classification of malignant tumors of the breast and female genital tract even does not include MFH in the sarcoma types that could grow in the breast[9]. This proves the rarity of this kind of tumour in the breast.

The origin of MFH is still controversial[10]-[12], with three existing theories dominating: (1) tissue cell origin; (2) fibroblast cell origin; and (3) primitive mesenchymal cell origin. The most common belief is that MFH is an intermediate type, and is derived from primitive mesenchymal cells that grow during the process of tissue cell and fibroblast cell differentiation.

According to the main morphological characteristics of tumor cells, MFH can be grouped into five types. First is the pleomorphic type, where the most prominent morphological features include storiform arrangement of the tumour cells along with multinucleated giant cells. Inflammatory cells in are occasionally mixed with the tumor cells. The second type is the myxoid type, where few of tumor cells are scattered in the loose myxoid matrix occupying more than 50% of the area. Third is inflammatory type where the most prominent feature is a large number of tumor cells in various differentiations mixed with a large number of inflammatory cells, particularly neutrophilic granulocytes. Fourth is the giant tumor cell type where the tumor is formed by the fibroblasts, histiocytes, and numerous osteoclast-type giant cells. Fifth is the angiomatoid type, characterised by both formation of lacunae as well as solid areas. In the present case the tumour seemed to be the pleomorphic type of MFH. For proper management of a patient differentiating malignant fibrous histiocytoma from other spindle cell lesions is very important. Differential diagnosis of MFH includes malignant phyllodes tumor and metaplastic carcinoma. Specific morphological features like biphasic architecture and epithelial component recognise the former. The latter is recognised by presence of a carcinomatous component or based on a cytokeratin immunopositivity of the neoplastic spindle cells. Presently both histopathological features like storiform pattern, tumour giant cells with atypical mitoses and immunohistochemical features support to diagnose our case as a genuine primary malignant fibrous histiocytoma of the breast. Our case is noteworthy, not only because of the infrequency of MFH in the breast, but also because of its de novo appearance and relative young age of the patient.

Surgery specially wide excision with tumor-free margins remains the mainstay of treatment. In contrast to breast carcinoma, role of axillary lymph node clearance is debatable. Radiation therapy and chemotherapy has also been used as adjunctive therapy in some cases [6]. For MFH, the role of chemotherapy in treatment is not entirely clear [13]. Radiation clearly reduces the incidence of local recurrence and has become an integral part of the treatment for MFH [14-15], especially where surgical margins are not widely free, as in our case but most important is the early detection which obviously has the greatest impact in the prognosis.

3. Discussion

After surgery the patient was treated with chest wall irradiation because of close deep margin. After few months she complained of upper limb weakness and pain. Then she underwent metastatic evaluation with computed tomography scan but no metastasis was detected. Thereafter she was referred to the Department of Physical Medicine and Rehabilitation for evaluation of her complaints where she was diagnosed to have postirradiation brachial plexopathy. Since then the patient is treated and followed up there.

4. Conclusion

Primary sarcoma of breast is a diagnosis of exclusion. It is important to differentiate this entity from similar looking tumours as they differ histogenetically as well as in biological behaviour highlighting the need of thorough sampling and the use of immunostain to arrive at diagnosis.

References


Author Profile

Dr. Tamalika Kundu got her MBBS and MD-Pathology degree from Burdwan Medical College and Hospital, Burdwan, West Bengal, India. Currently she is working as a senior resident cum demonstrator in ESI-PGIMSR & ESIC Medical College, Joka, Kolkata, India.